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Intrapancreatic accessory spleen: A case report and review of the literature

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ABSTRACT

We present the case of a 26 year old male who was found to have a mass in the tail of the pancreas on an ultrasound scan. The lesion was suspicious for a non-functioning pancreatic neuroendocrine tumour (PNET) and so he underwent distal pancreatectomy. Pathology revealed this to be an intrapancreatic accessory spleen (IPAS). This is a rare entity, and the literature on this subject is reviewed. A lesion in the pancreas that enhances in a manner similar to the spleen, whether the contrast is used in the setting of a Contrast Enhanced Ultrasound, a contrast enhanced CT scan, or a gadolinium enhanced MRI scan, is suggestive of IPAS. Nonetheless, the majority of these rare lesions are likely to be surgically excised rather than observed due to the similar appearance to PNET.

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1. Presentation of case

A 26 year old, otherwise healthy male presented with a month-long history of burning epigastric pain mostly in the evenings. The pain was not associated with food and was severe enough to keep him up at night. There were no associated fevers/chills, night sweats, nausea or vomiting. Gastroscopy was unimpressive except for some mild esophagitis and gastritis.

An ultrasound did not reveal gallstones or liver abnormality, however a homogeneous, well-defined, slightly hypoechoic mass was found in the tail of the pancreas (Fig. 1). MRI confirmed a well circumscribed, hypervascular, solid 1.4 cm lesion within the tail of the pancreas somewhat suggestive of an endocrine pancreatic lesion (Fig. 2). The patient was thoroughly questioned on signs and symptoms of a functioning pancreatic neuroendocrine tumour (PNET), however he denied all, including tachycardia, skin lesions, flushing, fainting, wheezing, and watery diarrhea.

Physical exam was unremarkable.

Endocrine work-up including C-Peptide, Gastrin, Glucagon, VIP and Ca²⁺ were all normal. Given the possibility this could be a

nonfunctioning PNET and after discussion with the patient, a distal pancreatectomy was planned.

Intraoperatively, palpation of the pancreas did not reveal an obvious mass therefore an intraoperative ultrasound was done, clearly showing a well encapsulated lesion in keeping with that found on the MRI. Spleen-preserving distal pancreatectomy was performed with the help of intraoperative ultrasound, to obtain a 1 cm margin. The postoperative course was unremarkable and he went home on the 7th day. His presenting symptoms resolved once he was started on ranitidine during the preoperative period.

The specimen received consisted of a piece of pancreas that measured approximately 8 cm × 5 cm × 4 cm. The cut sections revealed the presence of a well-circumscribed mass measuring 1.1 cm in diameter. The surrounding pancreas was unremarkable.

Microscopic examination of the mass showed the presence of masses of lymphoid tissue surrounding a central vessel (periarteriolar distribution) interspersed with complex network of venous sinuses indicated of splenic tissue (Fig. 3).

Immunohistochemical analysis with chromogranin, low molecular weight keratin, vimentin, CD45 and Masson delineated the non-pancreatic lineage of this nodule with strong CD45 positivity confirming its haemopoietic ancestry (Fig. 4).

Correlation of these pathological findings with the radiological images, confirmed the diagnosis to be that of an intrapancreatic accessory (normal spleen identified and confirmed on imaging) spleen (IPAS).

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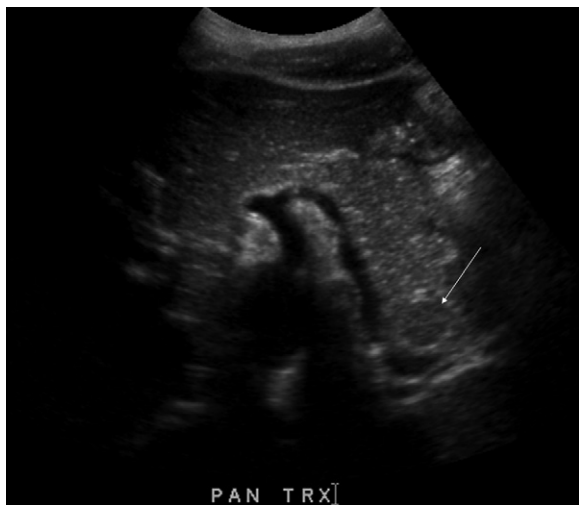


Fig. 1. Transverse ultrasound image showing a homogeneous, well-defined, round hypoechoic mass in the pancreatic tail.

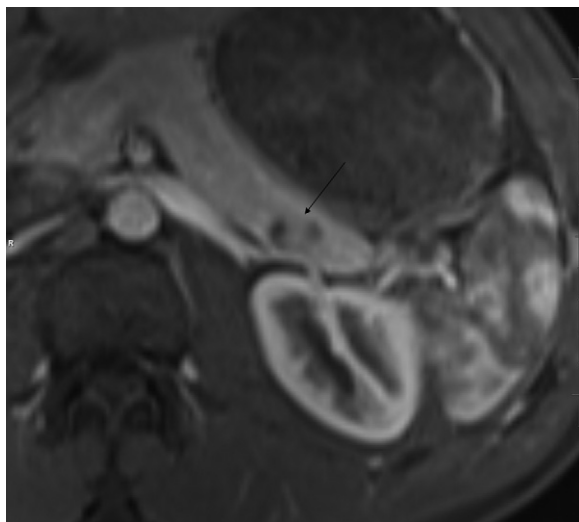


Fig. 2. Arterial phase post-gadolinium VIBE: note the broad band of enhancement through the center of the lesion, similar to the spleen.

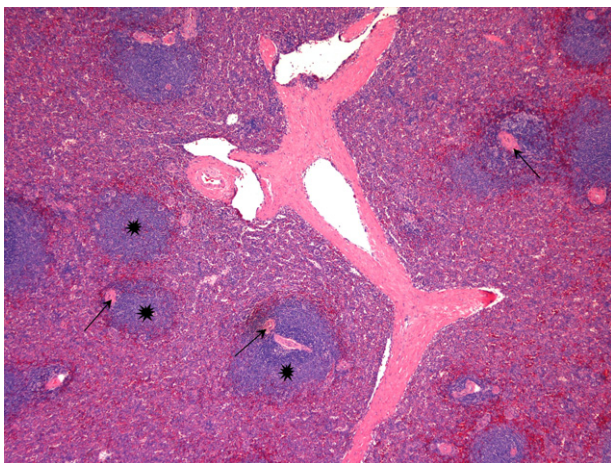


Fig. 3. This magnified image (2×) clearly confirms the histology of splenic tissue as demonstrated by the presence of lymphoid tissue (✱) surrounding a central arteriole (→).

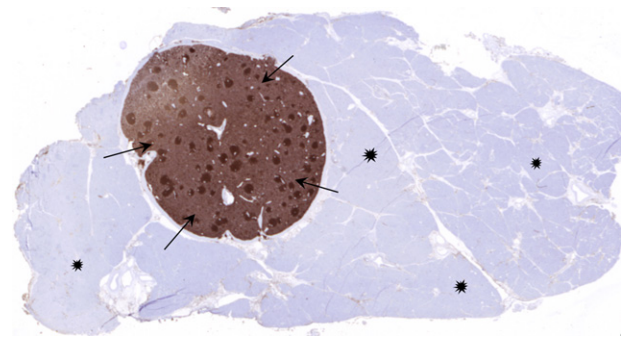


Fig. 4. Immunohistochemical staining with CD45 shows strong positive staining (→) of the mass confirming its haemopoietic ancestry with negative staining of the surrounding pancreatic tissue (✱).

2. Discussion

Intrapancreatic accessory spleen (IPAS) is a benign lesion related to an embryological aberration of splenic development. In a study of 3000 autopsies there were 364 accessory spleens (12.1%).¹ The majority (65%) of the accessory spleens were 1 cm in diameter or less. Accessory spleens are found most commonly in the hilum of the spleen or near the tail of the pancreas. Intrapancreatic accessory spleen is an uncommon entity with likely fewer than 30 cases reported in the literature.^{2–8}

As it is clinically silent, it is often discovered as a pancreatic mass in a patient being investigated for upper gastrointestinal symptoms. The preoperative workup is usually suggestive of a pancreatic neuroendocrine neoplasm. The correct diagnosis of IPAS is usually only confirmed after histopathological examination of the resected mass.⁴

Technetium-99m-HDRBC scintigraphy can identify a focal high concentration of red blood cells⁹ such as that seen in splenic tissue. However, the small size of most IPAS and the low resolution of scintigraphy may limit its usefulness in this setting.

Contrast-enhanced ultrasound may also be helpful in the diagnosis of IPAS. A study by Kim et al.,^{10,11} showed that in each of the four phases of contrast enhanced ultrasonography (CEUS) the IPAS demonstrates the exact enhancement patterns as visualized in the spleen.¹¹

When a mass is present in the pancreas on CT or MRI appearing as round or ovoid and well-demarcated, IPAS should be considered in the differential diagnosis.^{3,12} Perhaps one of the most striking findings suggesting an IPAS is that on a contrast-enhanced CT or MRI, the early arterial phase may demonstrate an inhomogeneous enhancement pattern similar to that seen in the spleen (Fig. 2). A PNET would be expected to enhance uniformly, even early in the arterial phase.

3. Conclusion

IPAS is an uncommon pancreatic lesion. A subtle lesion in the tail of the pancreas will most often be resected surgically for fear of missing a non-functioning PNET, a pancreatic adenocarcinoma, or a metastasis. However, certain features on contrast-enhanced CT or MRI or CEUS may be specific enough to allow for close follow-up rather than resection. No change in the lesion at 3 months would then provide more reassurance.

Conflict of interest statement

None.

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None.

Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor in Chief of this journal on request.

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